Complete aortic replacement in aortitis due to aseptic abscess syndrome

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ABSTRACT

A 36-year-old man was admitted for a tender inflammatory type IV thoracoabdominal aortic aneurysm with multiple aortic dilations. After open repair, he remained frail, but results of all infectious and inflammatory investigations were negative. Hypermetabolic intrasplenic collections were discovered on postoperative computed tomography, and aortitis with aseptic abscess syndrome was strongly suggested. Immunosuppressive therapy was undertaken, and his health improved dramatically. After 7 years of treatment, however, the initial aortic dilations had developed in size, necessitating multiple surgical procedures leading to complete aortic replacement. The postoperative course was uneventful with a satisfactory final computed tomography scan. Subsequent to immunotherapy, no new aneurysm developed. (J Vasc Surg Cases and Innovative Techniques 2020;6:216-20.)

Keywords: Aortitis; Complete aortic replacement; Thoraflex; FEVAR

Aseptic abscess syndrome (AAS) is a rare but severe disease often associated with inflammatory bowel disease (IBD) for which there exist few published data.[1] We report the case of an AAS involving multiple aortic aneurysms and requiring staged surgical procedures, leading to complete thoracic and abdominal aortic repair. To our knowledge, this is the first report of an association of noninfectious, nonatheromatous aortitis with AAS.[2] The patient signed a consent form authorizing us to publish the details of his hospitalization, images included.

CASE REPORT

A 36-year-old man was admitted in emergency for abdominal pain and fever. His history was remarkable for an isolated groin abscess surgically treated the year before. Enhanced computed tomography (CT) revealed multiple aneurysms with perivascular infiltration (Fig 1), including a 55-mm-diameter Crawford extent IV thoracoabdominal aortic aneurysm as well as other aneurysms of the aortic arch (47 mm in diameter), the descending thoracic aorta (36 mm), the left common carotid artery (15 mm), and the hypogastric arteries (25 mm on each side). At this stage, we found no family history of an aneurysmal disease. Surgical management of the thoracoabdominal aortic aneurysm was indicated, and the patient received an open aortobi-iliac bypass by thoracophrenolumbotomy. A beveled proximal anastomosis revascularized the visceral arteries with the exception of the left renal artery, which was separately reimplanted in the aortic graft. Given the high suspicion of infective aortitis, a cryopreserved arterial allograft was used. Probabilistic antibiotic therapy was immediately started after multiple samples of inflammatory tissues had been sent for bacteriologic and histologic analysis. The results of these different investigations, blood cultures, and serologic tests for bacteria and fungi were all negative, and the perioperative course was uneventful. The patient was discharged from the hospital, and the antibiotic treatment was discontinued. At 6 months, CT scan showed a patent bypass without any abnormality. The patient remained asymptomatic for a period of 12 months, after which he was once again admitted in emergency for rupture of the left iliac anastomosis, which required an emergent reintervention with an open prosthetic bypass. The postoperative period was uneventful. Once again, results of screening tests for infection and extensive laboratory tests for infectious diseases were negative, and blood cultures were sterile. The angiotensin-converting enzyme level and immunoglobulin G4 level were within normal ranges. Bone marrow biopsy revealed a reactive lymphoid hyperplasia, whereas a liver biopsy showed nonspecific mild hepatitis without infectious agents (including tuberculosis). Histologic evaluation highlighted a nonspecific cellular inflammation with aortic fibrosis associated with small granulomatous lesions at the intima-media junction and spumous cells with myxoid material in the intimal layer. Postoperative CT scan showed splenomegaly with intrasplenic collections (Fig 2), and positron emission tomography scan revealed hypermetabolism of the lymph nodes and spleen.

At this stage, inflammatory diseases such as Takayasu, Behçet, Whipple, Sweet, and Erdheim-Chester were ruled out.[3]

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Author conflict of interest: none.

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The editors and reviewers of this article have no relevant financial relationships to disclose per the Journal policy that requires reviewers to decline review of any manuscript for which they may have a conflict of interest.

2468-4287

https://doi.org/10.1016/j.jvscit.2020.02.011

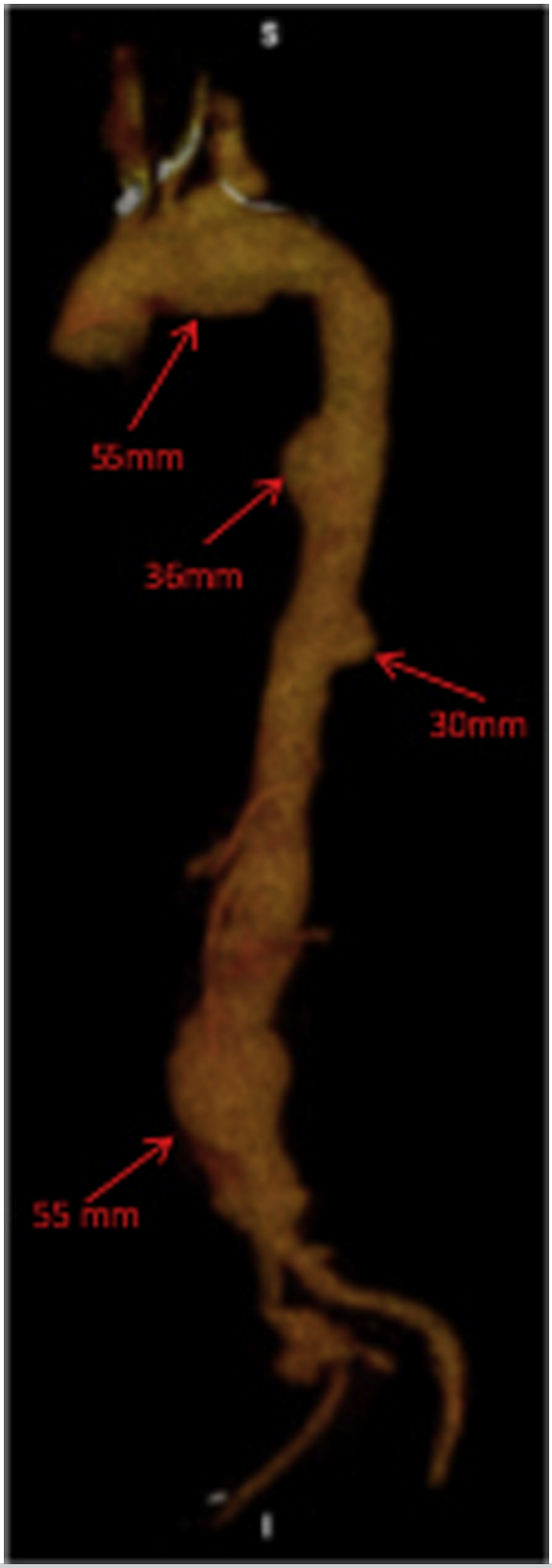

Fig 1. Initial computed tomography (CT) scan with symptomatic inflammatory 55-mm type IV thoracoabdominal aortic aneurysm and asymptomatic aneurysms of the aortic arch (47 mm) and the descending thoracic aorta (36 mm and 30 mm).

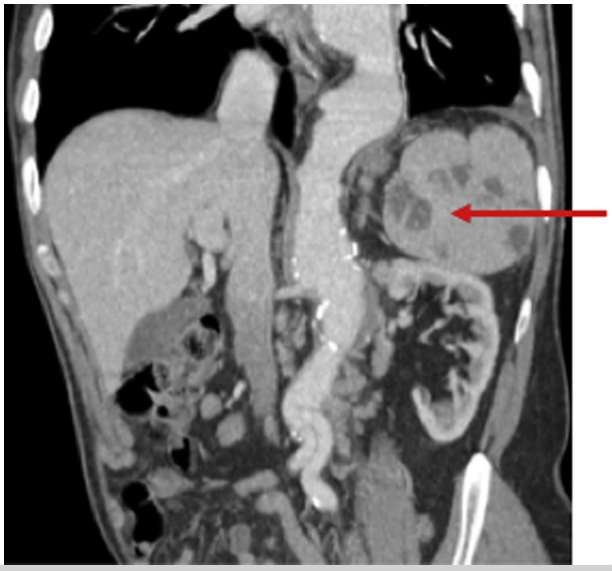

Fig 2. Computed tomography (CT) scan showing splenomegaly and multiple splenic abscesses (*arrow*).

Searches for Crohn disease and ulcerative colitis were negative. Oral corticosteroids were introduced in association with azathioprine and subsequently replaced by cyclophosphamide. Whereas splenectomy was initially considered, after discussion with internal medicine specialists, it did not appear essential to confirmation of AAS diagnosis. Furthermore, although it is possibly effective in management of other hematologic diseases, splenectomy did not seem to improve the patient's prognosis, and spleen rupture has never been reported in AAS patients.

In any event, the treatment was not effective, as shown by persistent fever, elevated C-reactive protein concentration (100 mg/dL), and leukocytosis (15,000/mm^3). Moreover, given the patient's incomplete adherence to treatment, it was decided to switch to monthly infliximab injections. The abscesses then regressed, and the patient was discharged home. Although follow-up was uneventful for 7 years, AAS was suggested because of the association of deep abscesses with neutrophilic features, negative results of screening for infection, and failure of antibiotic therapy, whereas prompt clinical and radiologic improvement was achieved with immunosuppressive therapy.[4]

Eight years after the first operation, the right hypogastric aneurysm had increased to 40 mm in diameter and was percutaneously embolized. By that time, the aortic arch aneurysm had reached a diameter of 60 mm (Fig 3, *A*) and necessitated an open frozen elephant trunk procedure with a Thoraflex hybrid prosthesis[5] (Vascutek, Terumo, Inchinnan, United Kingdom; Fig 3, *B*). One year later, the diameters of the thoracic aneurysms had increased from 36 to 60 mm and from 30 to 47 mm, respectively. Extension of the previous Thoraflex stent graft by two endovascular aortic repair procedures was carried out within a 6-week delay to decrease the risk of paraplegia. A few weeks later, the left hypogastric aneurysm (60 mm in diameter) was percutaneously embolized and the left carotid aneurysm

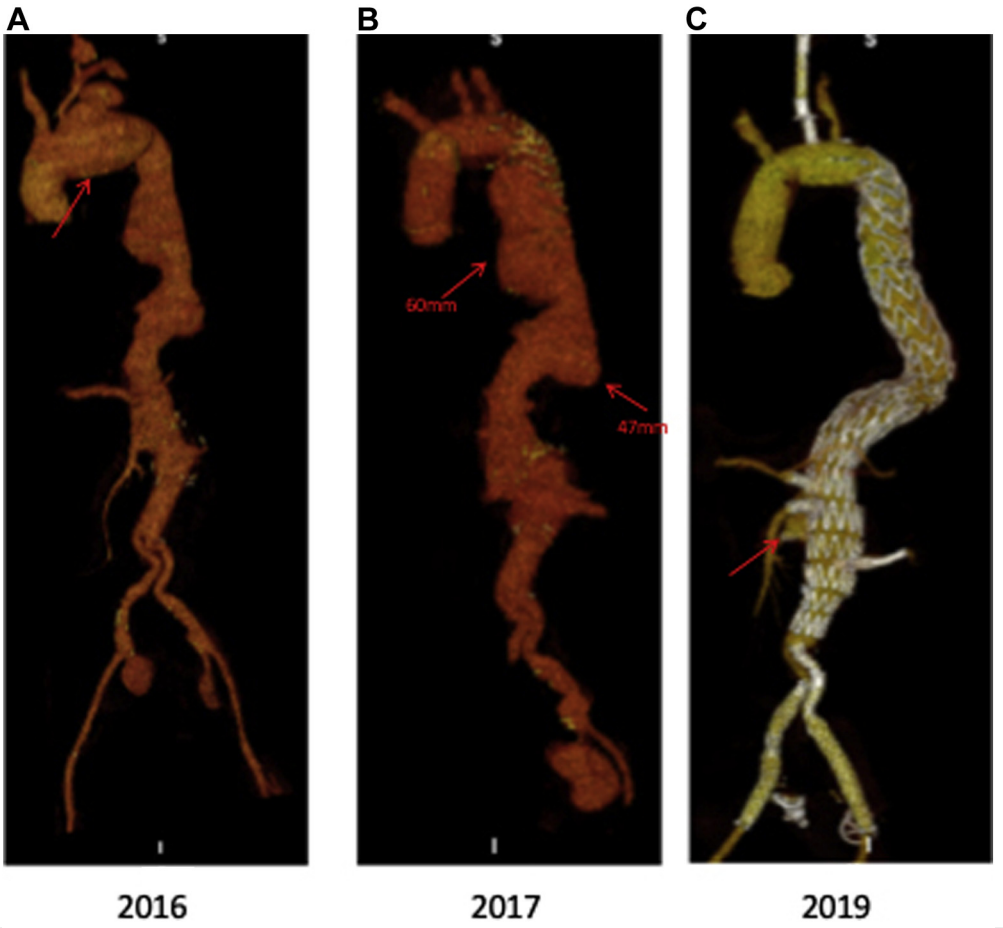

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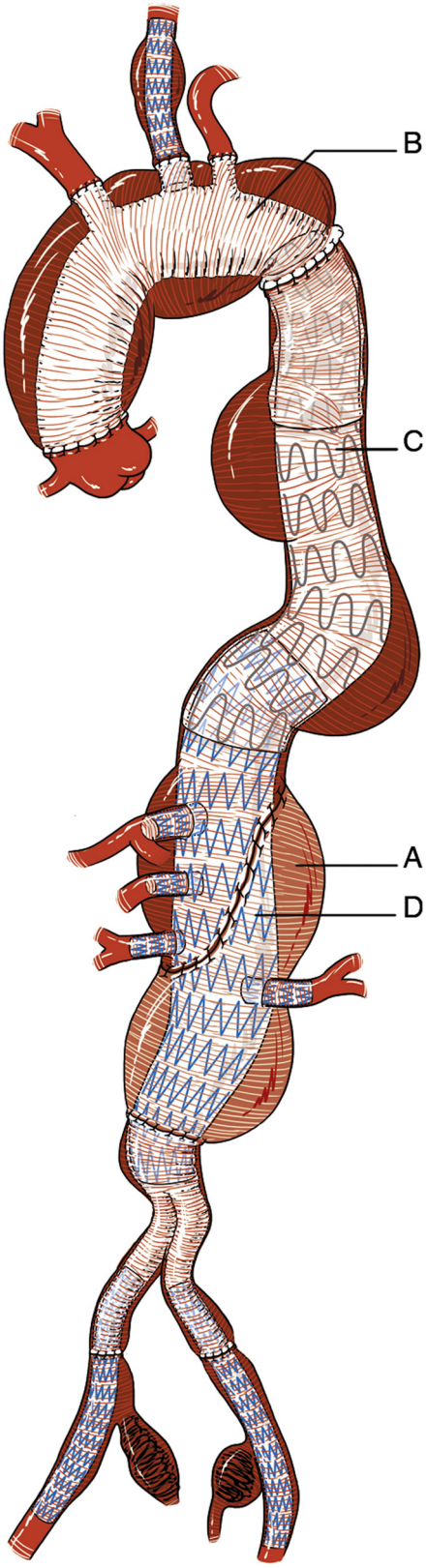
Fig 3. A, Aortic arch aneurysm grows to 60 mm (2016). **B,** Intermediate computed tomography (CT) scan after Thoraflex procedure showing an evolution of both descending thoracic aortic aneurysms to 60 mm and 47 mm (2017). Beveled proximal allograft anastomosis has already started to dilate. **C,** Final CT scan (2019) after thoracic endovascular aortic repair and four-vessel fenestrated endovascular aneurysm repair and bihypogastric embolization, showing type III endoleak around the left renal artery (*arrow*).

(35 mm in diameter) was stented. The patient recovered satisfactorily from the different procedures.

Finally, 1 year later and 10 years after the initial surgery, we observed an aneurysmal growth up to 65 mm in diameter of the beveled allograft anastomosis. The patient underwent four-vessel fenestrated endovascular aneurysm repair and subsequently presented with paraparesis, which improved within a few weeks notwithstanding mild neurologic claudication. Three months after fenestrated endovascular aneurysm repair, the last CT scan showed a satisfactory outcome except for a type III endoleak at the level of the left renal stent (Fig 3, *C*), which, given the absence of aneurysmal growth, was left alone to allow the spinal cord to develop collateral arteries.[6] All of these procedures are summarized in Fig 4.

DISCUSSION

Although AAS is a rare condition, it normally should be recognized by a vascular surgeon examining a patient with suspected septic aortic aneurysm. AAS occurs in young adults with IBD or granulomatous diseases in their family history. Whereas IBD was excluded in this case, even normal findings on colonoscopy do not rule out the possibility of later occurrence. For example, in the series of André et al,[7] IBD occurred up to 4 years after the first abscess. Fever may precede AAS by several months, accompanied by abdominal pain and a heightened leukocyte count. As observed in this case, AAS has a predilection for lymphoid organs, such as lymph nodes and spleen. In a patient with an aortic aneurysm, occurrence of abscesses in the spleen with negative microbiologic cultures is evocative of AAS. In our patient, *Yersinia* infection and Whipple disease were eliminated, as were slow-growing bacteria such as *Chlamydia trachomatis*, nontuberculous mycobacteria, fungi, and parasites, possibly yielding the same pathologic findings. Behçet disease and giant cell arteritis were likewise eliminated. Although the theoretical possibility of an



unknown pathogen was not definitively excluded, the positive response to monoclonal antibody therapy rendered it unlikely.[7] If a cryopreserved aortic homograft was initially preferred, it was because we suspected an infectious aneurysmal disease. As described by other authors, rupture of the distal suture line may have been related to use of the homograft.[8]

The pathophysiologic mechanism of AAS remains unclear, and its association with the growth of aortic aneurysm had not been described previously. By analogy with skin diseases involving neutrophilic infiltration, three mechanisms leading to the development of AAS have been hypothesized: vasculitis, as in Behçet disease; T-cell-mediated neutrophilic activation, as in Sweet syndrome; and autoantibody and complement initiation.[9] In this patient with AAS, it was hypothesized that this extensive and evolutive aneurysmal disease was characteristic of systemic vasculitis.

CONCLUSIONS

To our knowledge, this is the first report on an association of AAS and aortitis with multiple aneurysms. Notwithstanding its rarity, this association should be recognized by physicians. More precisely, a diagnosis of AAS associated with an aortic aneurysm should be considered in a patient in whom deep abscesses develop with negative results of repeated and extensive searches for an infectious cause. As AAS is a diagnosis of exclusion, in relapsing patients, it should be considered insofar as only immunosuppressive therapy is likely to achieve stability.

Fig 4. Several procedures were required in this patient and resulted in a complete replacement of the aorta. In chronologic order: A type IV thoracoabdominal aortic aneurysm received a cryopreserved arterial allograft **(A)**. One year later, the patient presented with a distal rupture of the allograft managed by implantation of a prosthetic bypass between the allograft and the common iliac arteries. Eight years later, a frozen elephant trunk procedure with a Thoraflex hybrid device was implanted to treat a 60-mm aortic arch aneurysm **(B)**. Because of the increase in diameter of the descending thoracic aneurysms, thoracic endovascular aortic repair was completed with a proximal sealing zone located in the Thoraflex device **(C)**. Ten years after implantation of the allograft, proximal aneurysmal evolution of the beveled anastomosis led to the implantation of a four-branched stent graft between the thoracic stent graft and the aortobi-iliac prosthesis **(D)**. In addition, both aneurysms of the internal iliac arteries were embolized 2 years apart, and the growing aneurysm of the left common carotid artery was excluded by a carotid stent.

REFERENCES

1. Bollegala N, Khan R, Scaffidi MA, Al-Mazroui A, Tessolini J, Showler A, et al. Aseptic abscesses and inflammatory bowel disease: two cases and review of literature. Can J Gastroenterol Hepatol 2017;2017:5124354.
2. Stone JR, Bruneval P, Angelini A, Bartoloni G, Basso C, Batoroeva L, et al. Consensus statement on surgical pathology of the aorta from the Society for Cardiovascular Pathology and the Association for European Cardiovascular Pathology: I. Inflammatory diseases. Cardiovasc Pathol 2015;24:267-78.
3. Rousselin C, Pontana F, Puech P, Lambert M. Differential diagnosis of aortitis. Rev Med Interne 2016;37:256-63.
4. André M, Aumaître O. Aseptic abscesses syndrome. Rev Med Interne 2011;32:678-88.
5. Shrestha M, Bachet J, Bavaria J, Carrel TP, De Paulis R, Di Bartolomeo R, et al. Current status and recommendations for use of the frozen elephant trunk technique: a position paper by the Vascular Domain of EACTS. Eur J Cardiothorac Surg 2015;47:759-69.
6. Spanos K, Kölbel T, Kubitz JC, Wipper S, Konstantinou N, Heidemann F, et al. Risk of spinal cord ischemia after fenestrated or branched endovascular repair of complex aortic aneurysms. J Vasc Surg 2019;69:357-66.
7. André MF, Piette JC, Kémény JL, Ninet J, Jego P, Delèvaux I, et al; French Study Group on Aseptic Abscesses. Aseptic abscesses: a study of 30 patients with or without inflammatory bowel disease and review of the literature. Medicine (Baltimore) 2007;86:145-61.
8. Antonopoulos CN, Papakonstantinou NA, Hardy D, Lyden SP. Cryopreserved allografts for arterial reconstruction after aorto-iliac infection: a systematic review and meta-analysis. Eur J Vasc Endovasc Surg 2019;58:120-8.
9. von den Driesch P. Polymorphonuclears: structure, function, and mechanisms of involvement in skin diseases. Clin Dermatol 2000;18:233-44.

Submitted Jan 3, 2020; accepted Feb 20, 2020.